

ON

SCLEREMA & ŒDEMA NEONATORUM.

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SCLEREMA and œdema of the newborn infant are affections concerning which great confusion reigns not only in the minds of medical men, but also in a majority of the textbooks devoted to the description of the diseases of infancy and childhood. It is my wish in this communication in the first place to show the manner in which this confusion has arisen, and, secondly, to demonstrate that the two diseases are pathologically quite distinct from each other. I shall first give brief clinical notes of two cases which occurred at the Edinburgh Maternity Hospital three winters ago and which were examples, the one of sclerema, and the other of œdema neonatorum.

Case of Sclerema Neonatorum.—On December 27th, 1885, an unmarried woman of 19 years of age was delivered at the Edinburgh Maternity Hospital of a premature male infant. The mother was a primipara, and was in apparently perfectly good health. The labour was not a long one, the first stage lasting 12 hours, the second half an hour, and the third ten minutes. The vertex presented in the O.D.P. position. The child was small; it weighed only 4lbs. 12ozs., and had a length of 18 inches. The placenta, which was normal, weighed 1lb. 3ozs., and the cord was 23 inches in length. The child, at the time of birth, had a premature appearance, and its breathing was with difficulty established, and, even when established, was shallow and weak. On the second day after birth it was noticed that the skin on the thighs, back, and buttocks, was hard and tense, and had a dirty yellowish appearance. Pressure with the finger did not cause any pitting of the skin. The child took the breast very badly, and the cry was weak and whining. The scleremic condition of the skin had, on the third day, spread to the neck, chest, and limbs, and was indeed almost universal. The body of the child felt cold to the touch, resembling the sensation given by contact with a half frozen corpse. The child died on the evening of the third day after birth. Through the kindness of Professor A. R. Simpson I was enabled to make a *post-mortem* examination of this, and of the following case, and to the conditions found at the necropsies I shall again return.

Case of Œdema Neonatorum.—On January 4th, 1886, a married 3-para of 31 years of age was delivered in the Maternity of a premature male child weighing 6lbs. 3ozs. The labour, which lasted for 16 hours, was complex, for the mother was at the time suffering from acute bronchitis and had pneumonic patches in both lungs. The vertex presented in the O.L.A. position. The mother was removed to the Royal Infirmary fourteen days after labour and

there she made a tedious recovery. The child, which measured 18 inches in length, and had a wasted appearance, developed in a few hours after birth a markedly oedematous condition of the skin of the lower part of the trunk, of the external genital organs, and of the lower limbs, especially on their posterior aspect. The skin in these regions pitted readily on pressure, and had a bluish cyanotic appearance. The surface of the child's body felt cold, and the limbs hung limp and inert. The child was put in the incubator, friction was applied, and stimulants were given; but notwithstanding these therapeutic measures, death ensued two days after the birth. The child passed no urine. It may be stated that the placenta weighed 1lb. 6ozs., and that the cord measured 19 inches in length. Neither the placenta nor the cord showed any abnormality.

These two cases enabled Professor Simpson to demonstrate to the students in attendance the differences which existed in the symptomatology of the two diseases, and to these differences I shall make fuller reference in the course of this paper.

Historical Note.—The history of sclerema and of oedema neonatorum forms a curious chapter in the annals of medicine. Uzembesius in 1718 reported a case of what was no doubt sclerema, which was peculiar in being congenital, and which he attributed to maternal impressions, for the mother of the scleremic infant had, during her pregnancy, spent much time in the churches contemplating the marble statues therein! The most important early contribution to the subject of sclerema came from the pen of Underwood. This author gave a most correct and complete clinical picture of the disease, and indeed his description has since scarcely been excelled. The disease he described was true sclerema, an induration of the subcutaneous tissue. Andry read Underwood's paper at a time when he had under his care several cases of oedema neonatorum, and thinking that the disease with which he was acquainted was the same as that described by Underwood, he gave to it the name of "sclérème." Underwood pointed out this error in a later paper and described the differences between sclerema and oedema; but the confusion thus introduced was perpetuated by subsequent writers, and it was not till the time of Parrot that the matter was cleared up. Auvity, Chambon, Capuron, and Léger all confounded sclerema with oedema, and Denis did not make the confusion less by describing two varieties of induration, a serous and an adipose or concrete. Billard seems to have had a clear conception of oedema, but sclerema he regarded as a cadaveric phenomenon, or as a condition which supervened only in the last few minutes of life. Bouchut still further complicated matters by comparing sclerema to the adult skin disease scleroderma. Coley, West, and other writers in this country followed in the main the descriptions and mistakes of the French physicians, and this statement applies also to the majority of German authors. In 1873, Clementowski, of Moscow, threw some light upon the subject when he clearly distinguished between oedema and adipose induration. The latter condition he regarded as due to inanition of the newborn infant and not as a cadaveric phenomenon. To Parrot, however, belongs the credit of most clearly differentiating between sclerema and oedema, even if one may not be able to go so far with him as to state that sclerema is one of the symptoms of athrepsia, whilst athrepsia or infantile atrophy is only one of the complications of oedema neonatorum. Depaul in the *Dictionnaire Encyclopédique des Sciences Médicales* t. xiii, f. 675 gives a very complete summary of the contributions to our knowledge of the two diseases, and Henoch in his work on *Diseases of Children* distinguishes clearly between sclerema and

œdema; but in modern textbooks there is still great confusion concerning these two diseases, thus Lewis Smith in the recently published American *System of Obstetrics*, vol. ii. p. 761, evidently includes œdema under the description of scleroma, whilst Ashby and Wright in their *Textbook of Diseases of Children* (1889) do not even mention sclerema. A few contributions of value have appeared in recent medical literature, of which we may mention Dr. Stephen Mackenzie's cases of sclerema and œdema which he showed not many weeks ago to the Clinical Society of London, and Dr. Barrs's "Notes of a case of Sclerema Neonatorum" which appeared in the *BRITISH MEDICAL JOURNAL*, May 4th, 1889. In concluding this short historical sketch of the literature bearing upon sclerema and œdema, I may add that the diseases have had many names given to them. Amongst the appellations which have been bestowed at one time upon sclerema and at another upon œdema, are scleroma, sclerisma, scleremia, œdematia, adipose induration, concrete induration, induration of the cellular tissue, and the skinbound disease.

This multiplicity of names has tended still further to complicate an already too complicated subject. I shall now pass in review the morbid anatomy, clinical features, etiology, diagnosis, prognosis, and treatment of sclerema and of œdema, noting the resemblances which exist between the two diseases, and at the same time calling attention to the marked differences which enable us to diagnose the one from the other.

Definition.—It is difficult to give a satisfactory definition of either sclerema or œdema, and in the present state of our knowledge an entirely correct one is impossible.

Sclerema may be provisionally defined as a rare disease, occurring most commonly in the newborn infant, characterised by induration of the subcutaneous tissue, and being little amenable to treatment. Until the pathology of sclerema and the nature of the physiological processes underlying its pathology are more fully understood, a more scientific definition than that given above is, I think, impossible.

Edema neonatorum may be defined as a disease of the newborn infant, characterised by serous infiltration of the subcutaneous tissue, due in most cases to infantile cardiac, renal, or pulmonary disease. *Edema neonatorum* is, therefore, manifestly more truly a symptom of several diseases than a distinct pathological entity; but until we have a fuller acquaintance with the symptomatology of cardiac, renal, and pulmonary disease in the newborn, and until we are better able to diagnose one variety of œdema from another, we must be content to study under one designation the characteristic features common to all.

Morbid Anatomy.—I was able to make a *post-mortem* examination of the scleremic infant within twenty-four hours after death, and I then found, in addition to the conditions of the heart, vessels, and viscera peculiar to the newborn infant, the following pathological appearances. The lungs were in a state of partial atelectasis, their posterior and lower portions being undistended with air; the abdominal viscera, especially the spleen, liver, and kidneys, were markedly congested; and the brain and its membranes were also congested. The thymus gland appeared to be normal. It was in the skin and subcutaneous tissue that the most marked pathological changes were to be found. The skin over the back, shoulders, thighs, and to a less extent over nearly the whole of the body was firm and tense, could not be pinched up between the fingers, and could not be made to pit on pressure. On making a section of the skin and subcutaneous tissue with a

knife a sensation was conveyed to the hand like that which one gets when one cuts bacon rind; and, on looking at the cut surface, one noted that the subcutaneous cellular tissue had a peculiar white glistening aspect quite unlike the yellowish appearance of the subcutaneous adipose tissue in the healthy infant. No serous fluid could be expressed from the cut surface, and to the naked eye the part did not appear to be congested.

In figure 1 are represented the microscopic appearances of the skin and subcutaneous tissue from the back of the scleremic infant. The outstanding feature in this drawing is the presence of a large quantity of brightly stained connective tissue which forms a network in the meshes of which lie the fat corpuscles. This connective tissue is very abundant and subdivides the subcutaneous adipose tissue into numerous patches of varying size. Not only are the bands of connective tissue increased in number, but they

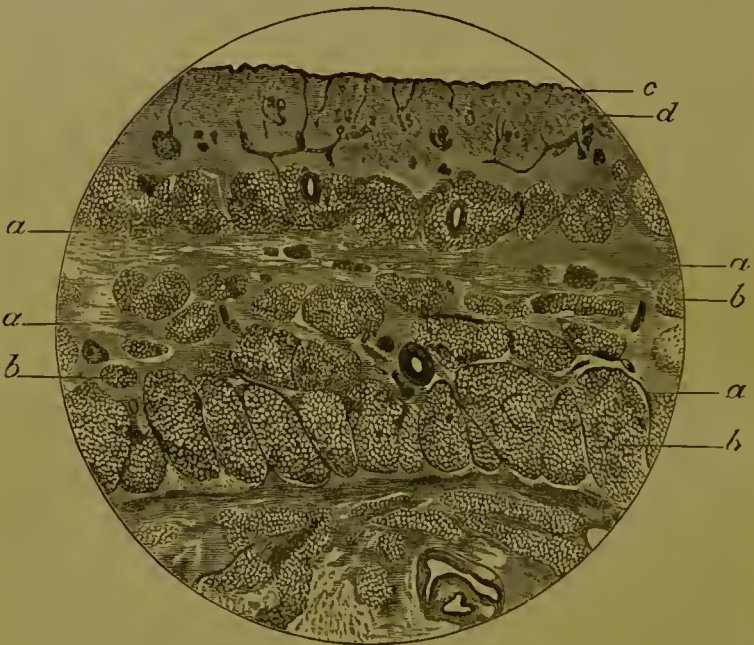


Fig. 1.—Sclerema neonatorum (section of skin of back $\times 18$). *a*, bands of connective tissue; *b*, clumps of atrophied fat cells; *c*, epidermis; *d*, rete Malpighii.

are also in many cases much thicker than is normal. In the true skin are seen the roots of a few hairs, and one or two sweat glands with slightly convoluted ducts. Such are the appearances noted with a magnifying power of eighteen diameters. Under a higher power it is seen that the fat cells have in some cases lost all their fat, and that in no case is the normal amount of oil present. The nucleus in all is clearly visible, and there is often also a rim of protoplasm underlying the cell wall. The cells making up the bands of connective tissue can be very clearly differentiated, and here and there are seen small vessels, surrounded by numbers of leucocytes, and pushing their way, as it were, into the clumps of fat cells. The papillæ are not well marked, and the outlines of the cells of the rete Malpighii are ill defined. The horny layer appears to be normal. The blood vessels in the papillæ are very small, but those in the adipose tissue are relatively large and

have, as has been noted, a small cell infiltration surrounding them. Parrot, whose description of the skin agrees in the main with that given above, considers the lesion to be a drying up of the skin with a consolidation of its layers and an atrophy of the adipose tissue. I believe that there is something more than a consolidation of the layers of the skin and atrophy of the fat cells, and am of opinion that there is an increase in the number and thickness of the connective tissue bundles, and that these bundles of fibres subdivide the masses of fat cells into smaller clumps and by pressure cause their atrophy. The starting point in the process is, I believe, the penetration of a capillary into a mass of fat cells, this capillary being accompanied by leucocytes which give rise to fibrous tissue which subdivides the mass of fat cells into smaller islands. The primal pathological factor may be, and probably is, a trophic lesion of the nervous system. This is a matter of hypothesis; but the

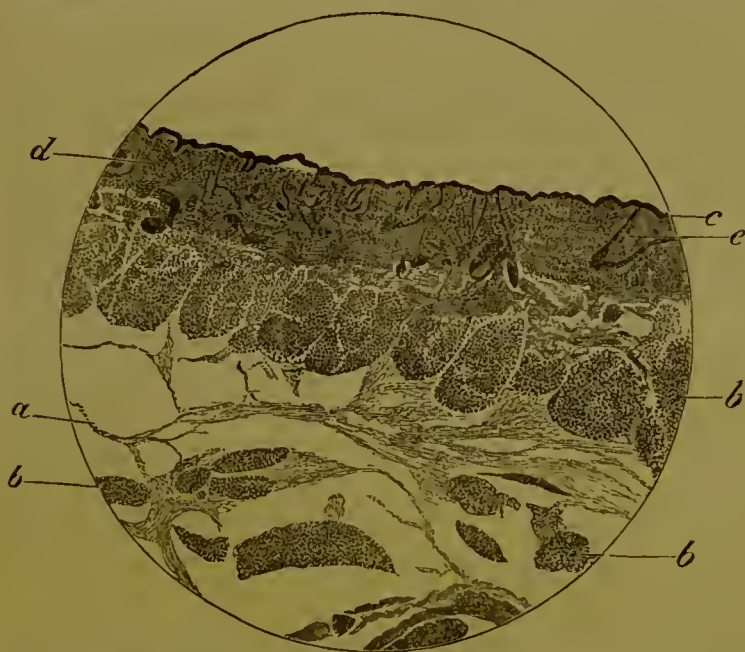


Fig. 2.—*Edema neonatorum* (section of skin of back $\times 18$). *a*, Loose bands of connective tissue separated by serous fluid; *b*, groups of atrophied fat cells; *c*, cuticle; *d*, rete Malpighii; *e*, hair.

hypothesis is strengthened by the fact that in three cases of sclerema reported by Dr. Angel Money¹, there was an accompanying paralysis, and by the resemblance which exists between sclerema and myxœdema.

The pathological appearances found in the infant with cedema differed in many respects from those which have been described in the sclerema case. Whilst the heart, liver, and spleen were normal, the kidneys showed on section an enormous degree of congestion, more especially of the cortex. The lungs, as in the sclerema case, were in a state of partial atelectasis. The skin of the back, abdomen, external genitals, and legs was of a bluish colour, had a soft feeling, and pitted readily on pressure. On making a section of the skin and subcutaneous tissue a somewhat congested surface was revealed, and a large quantity of watery

¹ *Lancet*, October 27th, 1888.

serum drained away from the tissue. The microscopic appearance of a section of the skin of the back is represented in figure 2. The masses of fat cells are seen to be held together in a loose manner by thin bands of connective tissue, and so loose is the connection that in many places the fat cells have fallen out of the section. It was very difficult to obtain a satisfactory section of the subcutaneous tissue in this case, a circumstance which contrasted very forcibly with the ease with which perfect sections of the skin, and underlying tissue were obtained in sclerema. In the oedema section the tissue is opened out from the presence of the serous fluid, whilst in the sclerema case the tissue is indurated, and the fat cells held in position by the hypertrophied connective tissue framework. The fat cells in oedema as in sclerema are atrophied, and the appearances of the cuticle and rete Malpighii are similar in the two diseases. The difference, then, appears to be in the subcutaneous adipose tissue layer.

The microscopic examination of the kidneys in the oedema case revealed the cause of the subcutaneous serous infiltration, for both kidneys were enormously engorged, the congestion being especially well marked in the cortex, and in both kidneys was there cloudy swelling of the cells of the tubules, and small cell infiltration of the Malpighian bodies. There was, therefore, evidence of the presence of tubular and glomerular nephritis, and this fact, taken in conjunction with the suppression of urine which was noted clinically, leads me to the conclusion that in this case of oedema neonatorum, the disease and the death were due to the kidney lesion. The infant died two days after birth, and the renal changes found might quite well have arisen in that time, so that I do not think there was here any foetal nephritis, at the same time the fact that the mother was suffering from bronchitis and pneumonia at the time of her confinement is noteworthy. No doubt there occur cases of oedema neonatorum in which the disease is due to lesions other than nephritis, such as cardiac and pulmonary diseases; but in this case the nephritic process was evidently the starting point of the pathological state. Oedema neonatorum I regard as a subcutaneous oedema, quite comparable to that which occurs in the adult and due to the same causes, whilst sclerema must, I think, be regarded as a disease peculiar to the newborn infant or young child, and most probably of the nature of a trophoneurosis.

Clinical Features.—In both sclerema neonatorum and oedema neonatorum the patients are weakly, often prematurely born, and in both diseases the body temperature rapidly falls below the normal. In sclerema the peculiar condition of the skin and subcutaneous tissue is found most markedly on the back, shoulders, and thighs, whilst in oedema the area of distribution corresponds with the lower part of the abdomen, the genital organs, and the back and legs. In sclerema the skin is firm and tense, cannot be raised in folds, and does not pit on pressure; in oedema, on the other hand, the skin is soft and boggy, can be pinched up between the fingers, and pits readily on pressure.

Etiology.—Very little is known concerning the etiology of sclerema. The disease occurs usually in prematurely born and weakly infants, which have been placed under bad hygienic conditions as to food, clothing, etc. It is, therefore, easy to understand how illegitimate foundlings are specially liable to it. It is most common in the winter months; but it is a rare disease in this country at any time of the year. It may occur in syphilitic infants, but any direct connection between the two diseases remains to be proved. With regard to oedema it may be said that

in many points the etiology is similar to that of sclerema. It occurs also in premature infants placed under bad hygienic conditions, and most probably cold has a powerful effect in leading to the renal, cardiac, and pulmonary states which are so often associated with it. It will be noted that both the cases recorded in this paper occurred during the winter months.

Diagnosis.—If attention be paid to the clinical features which have been enumerated, the diagnosis between sclerema and œdema ought not to be difficult. The possibility of sclerema being mistaken for tetanus neonatorum is not very great, for the rigidity of the limbs is never so great in the former disease, and the temperature curve in the latter disease will also serve to distinguish the two diseases.

œdema and erysipelas might possibly be mistaken for one another, and indeed œdema may occur during the course of erysipelas, but here also the condition of the temperature will serve as a diagnostic guide.

From syphilitic roseola œdema may be quickly differentiated by the result of antisyphilitic treatment.

It is possible that œdema neonatorum and general dropsy of the fœtus may be confounded; but the confusion is more likely to arise in medical literature than in medical practice. General dropsy is a disease of the fœtus and not of the infant, and the observations of Smith and Birmingham², seem to show that the disease is due to absence of the thoracic duct and of the lymphatics of the mesentery and skin. I saw one case of general fœtal dropsy three years ago, and, although I am unable to speak certainly with regard to the thoracic duct, yet the appearances of the skin and mesentery I find to agree entirely with those figured by Smith and Birmingham.

Prognosis.—The prognosis is very grave in both diseases, very few scleremic or œdemic infants recovering; but occasionally a case of recovery is reported in the medical papers, for example, Barrs's case in the JOURNAL, May 4th, 1889.

Treatment.—In both diseases the infant affected should be placed in an incubator. Friction with a stimulating liniment is indicated as is also "*gavage*." In cases of œdema where there is suppression of urine it would be well to try the effect of digitalis fomentations to the loins and of diuretics internally.

In conclusion, the hope may be expressed that with a fuller knowledge of the physiology and pathology of the newborn infant, these two diseases, at present so fatal, may become more amenable to treatment. At the same time it is well to remember that probably the diseased process is in both affections initiated *in utero*.

² *Journal of Anatomy and Physiology*, July, 1889.

